Dactylitis in a child with sickle cell trait

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Dactylitis commonly occurs in patients with homozygous hemoglobin S disease (sickle cell anemia), sickle cell–hemoglobin C disease or sickle cell–β-thalassemia. A case is reported of dactylitis associated with sickle cell trait, a very rare occurrence. It may be that in this patient the disorder was secondary to severe diarrhea and dehydration.

La dactylite est banale dans l'hémoglobinose S à l'état homozygote (la drépanocytose), l'hémoglobinose SC et la drépanocytose–β-thalassémie. On rapporte ici un cas très rare de dactylite chez une porteuse hétérozygote du gène de la drépanocytose. Une diarrhée sévère avec déshydratation a peut-être contribué à la pathogénèse.

Dactylitis, or hand–foot syndrome, is commonly the first clinical manifestation of sickle cell disease. Typically the child cries with pain, refuses to bear weight and has puffy, tender and warm feet or hands or both.[1-4] This phenomenon occurs in homozygous hemoglobin S disease (sickle cell anemia), sickle cell–hemoglobin C disease and sickle cell–β-thalassemia.[5-8] It has not previously been described in patients with sickle cell trait (heterozygous hemoglobin S disease). In this paper we describe a child with sickle cell trait who had dactylitis.

Case report

A previously well 14-month-old black girl was admitted to a community hospital with a 4-day history of nonbloody diarrhea and a 2-day history of fever. She was mildly dehydrated and irritable. Her rectal temperature was 40°C. The results of the remainder of the physical examination were unremarkable. Her diarrhea lessened with the institution of a clear-fluid diet, but she continued to be febrile. On the 4th hospital day her hands and feet became swollen. At that time she was transferred to our hospital for further evaluation.

The patient was alert but irritable, and she was in no acute distress. Her height, weight and head circumference were all at the 50th percentile. Her heart rate was 120 beats/min and regular, respiratory rate 22/min, systolic blood pressure 70 mm Hg and rectal temperature 39.5°C. The dorsum of her hands and feet was swollen, warm and tender to deep palpation. The remainder of the physical examination gave unremarkable results.

The hemoglobin level was 92 g/L, the hematocrit 0.28, the mean corpuscular volume 80 fL, the mean corpuscular hemoglobin level 332 g/L, the reticulocyte count 18 × 10^9/L and the leukocyte count 8.1 × 10^9/L (59% neutrophils, 37% lymphocytes and 4% monocytes). The erythrocyte sedimentation rate was 80 mm/h (Westergren method). The erythrocytes appeared normal except for a few poikilocytes and anisocytes. Specimens of blood, urine and stool were obtained for culture, and intravenous therapy with chloramphenicol and ampicillin was begun. No pathogens were cultured from the blood or the urine, but the stool culture yielded *Salmonella montevideo*. Hemoglobin electrophoresis revealed 56.5% hemoglobin A, 41.8% hemoglobin S and 1.7% hemoglobin A_2, a pattern compatible with a diagnosis of sickle cell trait. The child's father also had sickle cell trait, but her mother had normal results of hemoglobin electrophoresis. X-ray films of the child's hands and feet revealed soft-tissue swelling but no bony changes. A bone scan showed no abnormalities.

On the 3rd hospital day antibiotic therapy was stopped. The child became afebrile and appeared well clinically. The swelling in her feet disappeared on the 4th day. Her hands, however, remained swollen and minimally tender to palpation until the 7th day. She was discharged from hospital on the 9th day. At that time her erythrocyte sedimentation rate was 36 mm/h.

At follow-up 2 weeks after discharge physical examination revealed no abnormalities. X-ray films of the child's hands showed periosteal reaction of the fifth metacarpal in the left hand. X-ray films of her feet were normal. The hemoglobin level was 100 g/L, the hematocrit 0.30, the mean corpuscular volume 81 fL, the reticulocyte count 28 × 10^9/L, the leukocyte count 6.5 × 10^9/L, with a normal differential, and the erythrocyte sedimentation rate 15 mm/h.

Discussion

Dactylitis is the term used to describe painful, usually symmetric swelling of the dorsum of the hands and feet in children with sickle cell disease. It is presumably due to infarction of the metacarpals, metatarsals and phalanges, caused by sickling of the erythrocytes in the capillary beds with blockage of the vessels at many sites. It could also be the result of tissue anoxia secondary to capillary stasis caused by increased blood viscosity. The reparative response of hyperemia surrounding the infarcted area is manifested clinically by soft-tissue swelling and roentgenographically by the appearance of subperiosteal new bone 10 to 14 days after the swelling has subsided.[2] Thus, the condition is diagnosed clinically, with roentgenographic confirmation. This phenomenon occurs in homozygous hemoglobin S disease, sickle cell–hemoglobin C disease and sickle cell–β-thalassemia.[5-8]

Dactylitis is sometimes mistaken for osteomyelitis or Reiter's syndrome. Our patient could not have had osteomyelitis because her condi-

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tion improved despite only 3 days of antibiotic therapy. Interestingly, patients with homozygous hemoglobin S disease or sickle cell–hemoglobin C disease have a propensity for osteomyelitis secondary to infection with *Salmonella*.[4] Our patient showed no evidence of the conjunctivitis and urethritis associated with Reiter's syndrome.

Dactylitis has not previously been described in patients with sickle cell trait, presumably because sickling of erythrocytes in these patients is not common. Under certain extreme conditions, such as lack of appropriate pressure in an aircraft during a flight, infarction of the spleen has occurred in patients with sickle cell trait.[4] Why sickling would have occurred in our patient is not certain, but it could have been secondary to the stress of diarrhea and dehydration. Clearly dactylitis is unusual in patients with sickle cell trait and is much more likely to be due to sickle cell disease.

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